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SUMMER SUPPLEMENT

# Learning and sharing learning

No two patient encounters are alike. Patients may have different medical concerns. They may be of different gender, race, ethnicity, or socioeconomic status. They have different personalities, values, and life experiences. At the same time, healthcare providers will have a different perspective on each patient, based on their previous experiences and ongoing development. Consequently, each patient, situation, and context is unique, and the approach taken to each should be tailored accordingly.

Undoubtedly, this contributes to a dynamic work environment in medicine where every day presents new challenges, or at the very least, something different to do. Many people are drawn to medicine as a career for this reason, and to succeed, lifelong learning is critical. For medical students, classes may end in the spring, but learning never stops. Some pack their bags and remove themselves completely from the world of medicine, trekking deep into the wilderness, strolling along picturesque cobblestone roads, or exploring new cultures in a faraway land. Others shift gears and immerse themselves in research, delving into the questions that advance medical knowledge and understanding. And of course, others take medical learning into the real world with summer electives across the country and abroad. All of these experiences guide our personal and professional development, and by sharing them, these experiences enrich the lives and learning of those around us.

In the 2012 UWOMJ Summer Supplement, some of the summer clinical experiences of Schulich medical students are shared in the form of case reports. They provide insight into medicine, and the lives of patients and their caregivers. They also remind us that opportunities to learn are all around us, whether in the clinic or in our daily lives. I hope you enjoy reading this year's Summer Supplement, and are inspired to continue learning and sharing your experiences.

Jason L. Chan  
Junior Associate Editor

# An unusual case of constrictive pericarditis

Vidushi Khatri (Meds 2015)

Faculty Reviewer: Dr. Warren Teel, MD (Occupational Health Clinics for Ontario Workers, Sarnia-Lambton)

## CASE

A 61-year-old retired clerical worker, WM, presented to his family doctor in 2003 with a several month history of worsening exertional dyspnea and abdominal bloating. Previous medical history consisted of childhood pneumonia and a twenty-seven pack-year smoking history, having quit twenty years previously. His previous chest x-rays had shown bilateral pleural thickening and plaque formation, suggestive of prior asbestos exposure acquired while working for thirteen years in a foundry.

On examination, abdominal distension and ascites were noted. A thorough cardiac work-up was ordered, including a CT and echocardiogram. The CT revealed more extensive pleural plaques than shown in previous chest x-rays, with significant pleural effusions and some pleural calcification. The pericardium was also noted to be thickened. The echocardiogram with colour Doppler suggested the diagnosis of constrictive pericarditis (CP). A subsequent cardiac catheterization indicated significant coronary artery disease (75% stenosis in the left anterior descending artery) and a hemodynamic profile consistent with constrictive pericarditis. MRI identified pericardial thickening measuring 5 mm in thickness, along with pleural thickening and calcification.

Thoracentesis revealed that the pleural effusions were benign. As WM was feeling increasingly fatigued and short of breath by early 2004, he elected for surgery. He underwent a total pericardiectomy, however his LAD stenosis was not bypassed as his tissue was considered very friable. Subsequent pathology of the pericardial tissue revealed severe chronic inflammation and fibrosis consistent with constrictive pericarditis. WM had an immediate improvement in respiratory status and energy levels post-operatively. He submitted a claim to the Workplace Safety and Insurance Board of Ontario (WSIB) for compensation for asbestos exposure resulting in constrictive pericarditis. The claim was subsequently accepted and awarded by the WSIB in 2005.

## WHAT IS CONSTRICTIVE PERICARDITIS?

The pericardium is a fibroelastic sac that surrounds the heart. In constrictive pericarditis (CP), scarring and fibrotic changes alter the pericardium and prevent it and the heart from expanding.

The majority of constrictive pericarditis cases are idiopathic or post-cardiac surgery or post-radiation therapy in etiology.<sup>1-3</sup> In the developing world, post-infectious causes (i.e. tuberculosis or purulent pericarditis) are most common. A handful of cases – only 12 reported in literature<sup>4</sup> within the last twenty years – have been related to asbestos exposure.

## HOW DO WE RECOGNIZE CONSTRICTIVE PERICARDITIS?

As always, the history and physical exam play an important role in the diagnosis. Knowing that WM had pneumonia as a child may help estab-

lish associated artifacts that may appear on chest x-ray (e.g. granuloma from TB), but isolating the etiology would not help with definitive diagnosis or treatment in this case. CP patients often report symptoms of fluid overload and diminished cardiac output<sup>3</sup> in response to exertion. For instance, WM had been referred to a cardiologist when he experienced ascites, and his symptoms of shortness of breath and fatigue were present up until his surgery. On physical exam prior to his surgery, WM had only two of the six most common signs of CP,<sup>3</sup> which include elevated JVP, ascites, peripheral edema, pulsus paradoxus, Kussmaul's sign, and pericardial knock. His CT scan also revealed the fairly common CP finding of pleural effusions.

## HOW DO WE DIAGNOSE CONSTRICTIVE PERICARDITIS?

The initial steps of any cardiac work-up usually include EKG and chest x-ray. EKGs are not very useful for diagnosing CP, as there are no pathognomonic electrographic findings for CP.<sup>4</sup> Pericardial calcification may occasionally be caught on x-ray, however this is a rare finding.<sup>5</sup> Transthoracic echocardiography is the diagnostic test of choice for CP.<sup>6</sup> However, invasive hemodynamic evaluation during cardiac catheterization is often used to confirm the diagnosis, as was done with WM.

## WHAT IS THE TREATMENT FOR CONSTRICTIVE PERICARDITIS?

Although total pericardiectomy is the definitive treatment for chronic CP, diuretics can be used as a temporary measure. WM's abdominal ascites was controlled by diuretic therapy for several months until surgery. Total pericardiectomy is the treatment of choice, but partial pericardiectomy has also been conducted for some CP patients with prior asbestos exposure.<sup>7</sup>

## CONSTRICTIVE PERICARDITIS DUE TO ASBESTOS

WM is believed to be the first patient in Sarnia, Ontario to have developed CP in association with asbestos-related disease – one of only a handful reported in the medical literature. Without the concurrent development of pleural plaques, which are indicative of asbestos exposure, it is likely that the etiology of his CP would have remained idiopathic. Indeed, were it not for the increased prevalence of asbestos-related diseases in Sarnia, making both healthcare teams and city residents more aware of its consequences, the association of CP with asbestos exposure may have been missed altogether.

Several recent autopsy reports of workers at the Sarnia Occupational Health Clinic for Ontario Workers have described pericardial fibrosis with asbestos-related lung diseases and it is possible this finding is more prevalent than is realized. While asbestos-related pleural plaques will often increase physician's vigilance for asbestosis, lung cancer, and mesothelioma, it is worth considering the possibility of pericardial fibrosis and CP in workers with cardiorespiratory symptoms.

Although the medical literature has linked asbestos exposure with CP, major pieces of the puzzle have not been elucidated. It is unknown whether CP patients with prior asbestos exposure possess unique signs or symptoms that differentiate them from the majority of patients with CP. A more extensive case series could be conducted for clarification.

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# Acute unilateral rupture of the quadriceps tendon

Jimmy Yan (Meds 2015)

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## INTRODUCTION

Quadriceps tendon ruptures are relatively uncommon injuries.<sup>1</sup> It has been, however, chronicled through the ages; the earliest written report is attributed to Galen.<sup>2</sup>

While a rupture of the quadriceps tendon is a potentially severe affliction that requires accurate diagnosis and prompt management, it is an injury more frequently seen in older patients who are above 40 years of age.<sup>3-6</sup> The injury often follows an unexpected, rapid, high energy contraction of the quadriceps muscle, and can be associated with underlying pathological conditions of the quadriceps.<sup>3-6</sup>

## CASE

A 76 year old gentleman from the community of Ocean Falls, British Columbia, injured his right leg on July 1st, 2012, while jumping rope. He reported feeling and hearing a 'popping' in his right knee and he was no longer able to support his weight on that leg. An initial consultation at a local hospital included an ultrasound that was inconclusive and gave no clear diagnosis. The patient was given a GII knee brace to immobilize the knee for recovery. However, after several weeks there was little progress in his recovery.

He visited the Emergency Department of Vancouver General Hospital on August 8th, 2012, where the Orthopedics Trauma resident on call examined him. He was able to actively flex his right leg but could only passively extend it. He reported pain in the affected area during movement. When asked to contract the quadriceps muscle, the muscle bulk in the thigh was observed to be contracting, but no transference of force was observed through the lower leg. A suprapatellar gap was felt during examination, which is highly indicative of a complete tendon rupture. A lateral plain radiograph of the right knee was taken, which showed some avulsion fragments from the superior pole of the patella and a shadow across the soft-tissue in the region of the quadriceps tendon insertion into the patella (Figure 1).

The patient was admitted and scheduled for emergency surgical repair, which was handled by the Orthopedic Trauma team on-call on the following day. Regional anaesthesia was administered through a spinal block. A straight midline incision was made above the quadriceps tendon and knee. Surgical dissection and removal of hemarthrosis by suction revealed a complete quadriceps tendon tear at the osteotendinous junction with fibrotic tissue developing on the free edges of both quadriceps tendon and patella. Slight retraction of the muscle was noted but not measured. The fibrotic tissue was debrided and the tendon's anatomical insertion was roughened to create a fresh cancellous bone bed to facilitate healing. Two parallel suture anchors were drilled longitudinally into the superior pole of the patella. Using a Mayo needle, one end of each suture was sewn into the free end of the tendon. The edges were reap-

proximated and tied down proximally while the knee was in full extension. The reapproximated edges were reinforced with 1 Vicryl sutures and the Scuderi technique of using a partial-thickness flap of proximal tendon tissue folded over the rupture and sutured in place. The knee was taken through a 0° to 90° range-of-motion test to check the strength of the repair. 1 and 2-0 Vicryl were used to close the incision and surgical staples were used to close the skin. Post-operatively, the knee was immobilized in full extension.

## DISCUSSION

### *Anatomy and mechanisms of injury*

The quadriceps tendon is formed by the distal tendinous ends of all four muscles of the quadriceps: the rectus femoris, vastus intermedius, vastus lateralis, and vastus medialis.<sup>7</sup> These four muscles are innervated by the femoral nerve and coalesce into one band which attaches to the superior border of the patella bone.<sup>7</sup> Occasionally an anatomical variant of the articularis genu muscle can contribute fibers to the tendon.<sup>7</sup> The portion of the quadriceps tendon that inserts into the patella divides into three separate planes. The superficial, or anterior, plane is composed of the rectus femoris, while the second, or middle, plane contains both the vastus lateralis and medialis.<sup>7</sup> Finally, the deep plane consists of the vastus intermedius. Deep to all these planes is the synovium.<sup>7</sup>

The tendon is a component of the knee joint's extension system.<sup>4</sup> Connecting the patella to the quadriceps, it allows energy generated by those muscles during extension to be transferred through the patella and the



**Figure 1:** Lateral radiograph of R knee. Note avulsion fragments along the superior pole of patella, as well as a shadow in the soft-tissue region of the quadriceps tendon insertion into the patella.

patellar tendon into the tibial tubercle.<sup>4,6-7</sup> The quadriceps muscles can contract concentrically (with shortening of the muscles) or eccentrically (with lengthening of the muscles).<sup>4,5</sup> Much higher contractile forces can be generated eccentrically, and it is during this type of contraction, which occurs with a partially flexed knee and a planted foot, that tears typically happen.<sup>4,5</sup> These mechanics are often displayed in attempts to regain balance or landing while jumping,<sup>4,6</sup> which was how the injury manifested for our patient, as he reported to be jumping rope when he ruptured his tendon. Upon landing from a jump, the quadriceps experience a rapid and powerful eccentric contraction against the individual's full body weight.

It is important to note that the quadriceps tendon in a normal, healthy individual is an incredibly strong structure.<sup>8</sup> Past studies have found that this tendon can withstand remarkable stress loads without rupturing.<sup>1-2,8</sup> Tendons that rupture under lesser trauma loads are likely to have some form of pathological architectural change. These alterations can occur due to the result of degenerative changes, vascular disturbances, chronic conditions such as renal disease, uremia, diabetes, rheumatoid arthritis, hyperparathyroidism, lupus, gout, amyloidosis, and obesity, as well as steroid overuse.<sup>5-9</sup> Rupture due to direct trauma, laceration, or penetrating injury is rare.<sup>9</sup>

## Diagnosis

Ruptured quadriceps tendon clinically presents with a triad of knee pain, inability to actively extend the knee joint, and a suprapatellar gap.<sup>3-6,10</sup> The suprapatellar gap is considered pathognomonic for a complete tear,<sup>4</sup> however, it can be difficult to palpate due to edema secondary to hematoma or hemarthrosis.<sup>4,5,10</sup> Obstructing hematoma may have contributed to the missed diagnosis during the patient's first hospital visit.

Medical imaging can aid in the diagnosis. Lateral plain radiographs of the knee can reveal abnormalities such as a loss of the quadriceps tendon shadow, a suprapatellar mass, patella baja (an inferiorly displaced patella), calcified masses, joint effusion.<sup>11-12</sup> Ultrasound has a high sensitivity and specificity for determining the location of the tear and distinguishing partial and complete ruptures.<sup>13</sup> Ultrasounds are quick and non-invasive, but depend on operator skill. MRI has become the most consistent and accurate imaging modality for visualizing an injured quadriceps tendon.<sup>4,13</sup> It can be used in cases of extensive hematoma or edema, and is able to locate and depict the extent of injury.<sup>13</sup> It is useful for surgical planning; however, high costs reserve MRI use for when there is still doubt following all other diagnostic methods.<sup>13</sup>

## Treatment

Quadriceps tendon repair depends on the severity of injury. Partial tears do not require surgical treatment; the knee joint is immobilized in full extension for 6 weeks.<sup>3-4</sup> Subsequent physiotherapy for range-of-motion and quadriceps muscle re-strengthening are recommended.<sup>3-4</sup> Joint immobilization is gradually discontinued as the patient regains muscle control and discomfort decreases.<sup>3-4</sup> Hematoma or hemarthrosis drainage can promote healing.<sup>4</sup>

A completely ruptured quadriceps tendon requires surgical repair for optimal functional outcomes.<sup>14-15</sup> There are many methods that can offer favorable results, with no study designed to offer a comparison on which technique is the most efficient and reliable in delivering return of function.<sup>4,16-17</sup> Commonly, Krackow or Bunnell sutures are placed through the tendon using thick, non-absorbable sutures, which are subsequently passed through longitudinally drilled, parallel channels in the patella and tied distally.<sup>16</sup> Suture anchors drilled into the patella and tied to the tendon, which was the method used in this case, also yield good results.<sup>17</sup> Postoperative care involves keeping the knee joint immobilized in full extension with a hinged knee brace, as was done in this patient, for 4-6

weeks, followed by physiotherapy to regain range-of-motion.<sup>14-15</sup> The brace is typically removed after 12 weeks and good functional return should be achieved 12-16 weeks after the surgery.<sup>3-4,14-15</sup>

Delaying operation can lead to complications in the repair process as the quadriceps tendon begins to retract approximately 72 hours after injury.<sup>5,18-19</sup> There is some debate on treatment timeline, with some studies showing no detriment following a delay between injury and operation, and others showing worse results for delayed repairs.<sup>5,18-21</sup> In this case, there was an approximately 6 week delay between injury and repair, with noted muscle retraction during the operation. It remains to be seen how the patient's return to function fares following the repair.

This case report shows a relatively infrequent, but potentially debilitating, injury that can result from a seemingly harmless context. It emphasizes the importance of a thorough knee examination, as critical diagnostic clues can be easily missed. Once diagnosed, prompt surgical repair is recommended and a multidisciplinary approach to management is recommended to achieve optimal functional outcomes.

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## CASE REPORT

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# Sudden cardiac arrest in a young professional athlete

Jeffrey M. Landreville (Meds 2014)

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**C**ardiac arrest is an acute medical emergency characterized by the absence of cardiac activity, circulatory failure, abnormal respiration, and unresponsiveness. Historically, the term sudden cardiac death (SCD) has been used to describe all cardiac arrests. Current guidelines suggest that the label SCD should only be applied when attempts at resuscitation have failed and death occurs.<sup>1</sup> The term sudden cardiac arrest (SCA) is now used to describe those individuals whose cardiac activity has returned either spontaneously or following interventions such as cardio-pulmonary resuscitation (CPR) and/or defibrillation.<sup>1</sup>

Cardiac arrest is a common occurrence throughout the world. In Canada, a cardiac arrest occurs every 12 minutes with a yearly incidence of approximately 45,000.<sup>2</sup> Cardiac arrest can be thought of as an end event of any of the following four conditions: ventricular tachycardia, pulseless electrical activity, ventricular fibrillation, and asystole.<sup>3</sup> The causes by which these conditions/arrhythmias are generated can be broadly classified into those associated with overt structural heart disease (diagnosed on coronary angiography, echocardiography or resting electrocardiogram (ECG)) and those without (Table 1).<sup>4</sup> In general, ischemic heart disease is the most common cause of cardiac arrest, although less common causes are found more frequently in younger patients.<sup>5</sup> This case will highlight a rare cause of SCA in a young professional athlete.

## CASE

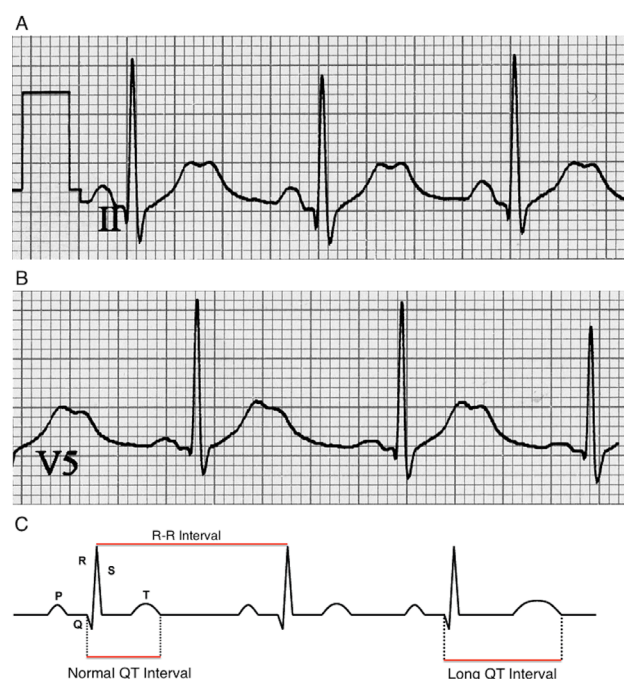
Mr. C is a previously healthy 25-year old professional hockey player who presented to hospital following SCA while swimming with friends. At the time of his arrest, he received CPR and was defibrillated three times by an automated external defibrillator for presumed ventricular fibrillation or ventricular tachycardia. Upon admission, Mr. C was intubated and transferred to the intensive care unit (ICU) where he underwent therapeutic hypothermia.<sup>6</sup> Following several days in the ICU, Mr. C was transferred to the coronary care unit (CCU) for further investigations and management.

Once in the CCU, Mr. C remained stable and the efforts of the medical team turned towards investigating the etiology of Mr. C's cardiac arrest. An echocardiogram revealed a normal left ventricular ejection fraction and mildly decreased right ventricular function with moderate dilatation of the right ventricle. Cardiac magnetic resonance imaging found no evidence of myocardial infarction, fibrosis, scarring, or findings suggestive of arrhythmogenic right ventricular cardiomyopathy. Coronary angiography showed normal anatomy with no obstructive coronary disease. Interestingly, while resting ECGs appeared normal, subsequent stress ECG testing revealed an abnormal QT interval. Genetic testing confirmed the diagnosis of congenital long QT syndrome (LQTS) type 1. Prior to discharge, Mr. C received an implantable cardioverter-defibrillator device (ICD). The remainder of his convalescence was unre-

markable and he will be followed by the electrophysiology service.

## DISCUSSION

First described in the 1957, our understanding of LQTS has grown enormously with the advancement of molecular biology and genetics.<sup>7</sup> LQTS is a rare disorder characterized by delayed myocardial repolarization that predisposes these individuals to a form of polymorphic ventricular tachycardia known as torsades de pointes.<sup>8</sup> This dangerous arrhythmia can quickly develop into ventricular fibrillation and subsequent cardiac arrest. The pathophysiology of LQTS relates to mutations in the genes responsible for producing the cardiac ion-channel proteins that regulate the flux of sodium and potassium ions during myocardial contraction.<sup>9</sup> To date, more than 150 different mutations in 7 genes have been implicated in LQTS.<sup>10</sup> The location and type of mutation, as well



**Figure 1.** Long-QT syndrome ECGs and measurement of the QT interval. A. Magnified lead II of an ECG taken during recovery following an exercise test. B. Magnified lead V5 of an ECG taken during recovery following an exercise test. C. The absolute QT interval is measured from the beginning of the QRS complex to the end of the T-wave. The R-R interval is measured from one R wave to the next R wave. The corrected QT interval ( $QTc = QT / \sqrt{R-R}$  [seconds]).

## CASE REPORT

**Table 1: Major causes of cardiac arrest**

Cardiac arrest with overt structural heart disease	Coronary Disease	Ischemic heart disease Anomalous coronary circulation
	Cardiomyopathies	Ischemic cardiomyopathy Hypertrophic cardiomyopathy Arrhythmogenic right ventricular cardiomyopathy
	Other	Wolff-Parkinson-White syndrome
Cardiac arrest without overt structural heart disease	Primary electric	Long-QT syndromes Short-QT syndromes Brugada syndromes
	Metabolic imbalance	Hyperkalemia/hypokalemia Hypocalcemia Acidosis
	Noncardiac	Pulmonary embolus Intracranial hemorrhage Drug induced

as variable genetic penetrance, help to explain the broad range of clinical presentations that are observed in this syndrome.<sup>9</sup>

The diagnosis of LQTS is based on history, 12-lead ECG, exercise and sympathomimetic drug provocation, and genetic testing. On history, patients may report palpitations, syncope, seizures, or no symptoms at all. It is important to inquire about a family history of cardiac arrest as well as several gene-specific symptom triggers including exercise, emotional states, loud noise, and sleep related events.<sup>11</sup>

The 12-lead ECG is an important tool in the diagnosis of LQTS (Figure 1). When interpreting the QT interval, it must be corrected for heart rate. This is achieved through the use of Bazett's formula ( $QT_c = QT / \sqrt{R-R}$  [seconds]) which is most accurate for heart rates between 60-100bpm.<sup>12</sup> Corrected QT intervals >440ms in males and >460ms in females are considered abnormally long.<sup>4</sup> Particular attention should also be given to the presence of characteristic T-wave morphology which has shown to be associated with certain types of LQTS.<sup>13</sup> The presence of a normal resting ECG does not exclude the diagnosis of LQTS. As exemplified in this case report, exercise or drug provocation may be required to bring out repolarization abnormalities in individuals who exhibit normal QT intervals at rest.<sup>14</sup>

The management of LQTS is complex and multi-faceted. Patient education, genetic counseling, and family screening are important to consider when caring for these patients. With regards to medical therapy, beta-blockers are first-line medications as they have been shown to reduce cardiac events.<sup>15</sup> In high risk individuals such as cardiac arrest survivors or those refractory to beta-blocker therapy, more aggressive management is indicated. Sympathetic blockade via left cardiac denervation surgery or ICD implantation should be strongly considered in high risk individuals.<sup>16</sup>

In summary, establishing a definitive diagnosis following SCA in the absence of overt structural heart disease remains challenging. The recommended approach is to obtain a comprehensive clinical history and employ sequential non-invasive and invasive investigations including provocative testing and advanced imaging techniques. Unfortunately, despite these efforts, the underlying cause in nearly half of these patients will remain unknown and a diagnosis of idiopathic ventricular fibrillation will be given.<sup>17</sup>

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# Gait disturbances and seizure-like episodes in a patient with a Chiari malformation

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## CHIARI MALFORMATIONS

Chiari malformations are a diverse group of congenital malformations that involve the cerebellum, brainstem, and craniocervical junction in the posterior fossa.<sup>1</sup> Chiari malformations were first described by John Cleland in 1883, but the term is used in recognition of Hans Chiari, who described a case of tonsillar ectopia in 1891.<sup>2</sup> The term Arnold-Chiari is also used to describe such malformations with spinal dysraphism, in recognition of Julius Arnold.<sup>2</sup> Three types of Chiari malformations have been identified to be compatible with life, each involving distinct clinical and anatomical features, but all involving various degrees of cerebellar descent through the foramen magnum.<sup>3</sup> Chiari malformations are diagnosed on the basis of neuroanatomical findings alone, and thus magnetic resonance imaging (MRI) is the primary modality for evaluation.<sup>4</sup> There are no tissue, blood or cerebrospinal fluid (CSF) biomarkers to confirm the diagnosis.<sup>1</sup>

The Chiari I malformation (CMI) is characterized by downward displacement of the cerebellar tonsils through the foramen magnum into the upper cervical canal.<sup>5</sup> Tonsillar ectopia of 5 mm or greater is generally consistent with CMI. However, an association between the degree of cerebellar descent and clinical severity is unclear.<sup>1</sup> As such, the clinical manifestations of CMI are highly variable, and may be due to the amount of nervous tissue displacement and the degree of compression.<sup>6</sup> Primary symptoms include tussive headaches, visual changes, syncope, muscle weakness, dysarthria and ataxia.<sup>7</sup>

Chiari II (CMII) and III (CMIII) malformations are associated with neural tube closure defects, and occur much less frequently than CMI. CMII is characterized by a small posterior fossa with downward displacement of the cerebellar tonsils and inferior cerebellar vermis into the foramen magnum and upper cervical canal, in association with a myelomeningocele.<sup>1-8</sup> These patients tend to have hydrocephalus,<sup>9</sup> and clinical manifestations may include lower cranial nerve deficits, cerebellar dysfunction, and respiratory disturbances.<sup>10</sup> CMIII is exceedingly rare, and is defined as cerebellar displacement into an occipital or cervical cephalocele.<sup>8</sup> This condition has a high early mortality rate, and causes several severe neurological deficits in survivors, including delayed milestones, seizures, ataxia, and spasticity.<sup>9</sup>

## CASE

LG is a 2 year old boy who presented in May 2012 with progressive gait ataxia and generalized weakness, in addition to falling down approximately six times per day. These symptoms were accompanied by slurring of speech, neck pain, and headache without nausea or vomiting.

His past medical history is significant for similar episodes, described as “grey spells,” which presented within the first few months of life. He has had multiple Emergency Department admissions for issues of fall-

ing, associated with respiratory symptoms and hypotonia. In addition, his mother would describe “shaking episodes” reminiscent of generalized tonic-clonic seizures. These symptoms were evaluated carefully: a thorough cardiac examination did not reveal any pathology, nor did an ophthalmological evaluation or a detailed genetic/metabolic screen (including blood and urine amino acids, urine organic acids, plasma ammonia, lactate, and carnitine levels, and urinary acylcarnitine profiles). Multiple EEG evaluations, including several 24-hour video EEGs revealed no evidence to support a diagnosis of clinical or electrographic seizures. A tentative diagnosis of alternating hemiplegia of childhood was made, which is a poorly understood disorder, and primarily a diagnosis of exclusion.<sup>11</sup>

MRI of the head demonstrated extension of the cerebellar tonsils 7 mm below the foramen magnum, which is consistent with CMI. At that time, it was decided to forego surgical intervention due to the moderate severity of his symptoms. However, he subsequently developed more frequent seizure-like activity without any supportive evidence of the events being epileptic in origin, and follow-up MRI demonstrated an additional extension of 4 mm. Thus, it was decided to perform a suboccipital craniectomy for posterior fossa decompression. Unfortunately, this surgery was complicated by two subsequent CSF leaks from the initial incision, as well as bacterial meningitis secondary to the CSF leak.

Following these complications, LG experienced only a single event prior to his current admission. While eating in a restaurant, his head flopped backwards, accompanied by a transient depression of his level of consciousness and abnormal extensor posturing. A thorough assessment revealed no focal neurological deficits, unremarkable physical exam and blood work, unremarkable EEG and MRI, and negative lumbar puncture cultures.

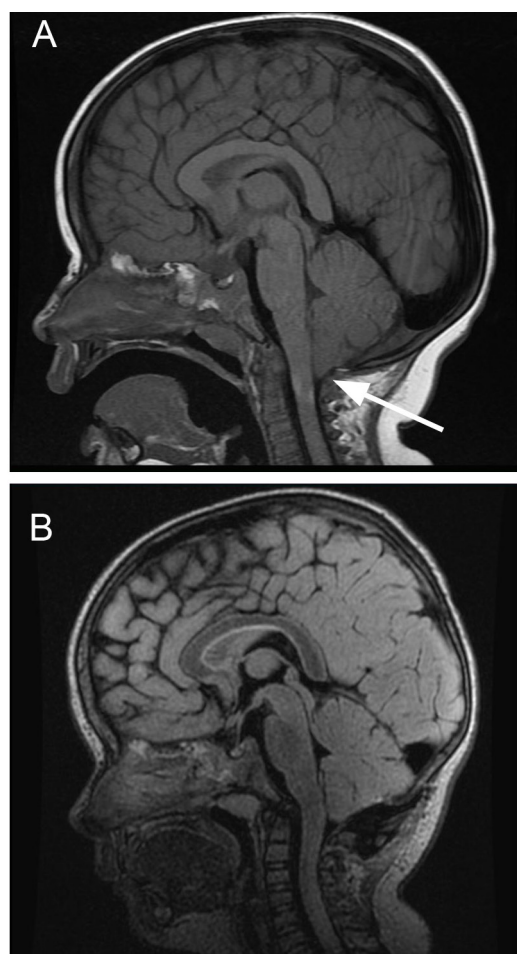
With regards to his current admission, MRI did not demonstrate any interval change. EEG evaluation again did not reveal epileptiform activity. LG did not demonstrate any symptoms in hospital as had been reported by his family, and he was discharged home without any additional therapy or medications.

## DISCUSSION

The presenting symptoms in patients with CMI are diverse and usually necessitate investigations for other pathologies before the definitive diagnosis is made. These manifestations can often be related to dysfunction of the brainstem, cranial nerves, spinal cord, or cerebellum.

The management of CMI depends on the presenting symptoms and associated neurologic impairments. It has been suggested that asymptomatic patients can be managed conservatively with serial MRI and regular clinic visits,<sup>12</sup> although some would advocate for prophylactic surgery to prevent neurological sequela.<sup>13</sup> The goal of decompressive surgery is to





**Figure 1.** Sagittal MR imaging demonstrating pre-operative cerebellar tonsillar herniation (A; T1) and subsequent decompression following suboccipital craniectomy (B; FLAIR).

restore CSF flow, and is indicated for symptomatic relief of syringomyelia, myelopathy, cerebellar symptoms, neck pain, or headache.<sup>9</sup>

In the case of LG, his symptoms led to a thorough neurological, cardiac, metabolic, and ophthalmologic work-up prior to diagnosis and treatment. Previous reports have identified a similar cohort of children who presented with drop attacks, extensor posturing, or varying degrees of respiratory compromise and described these episodes as “cerebellar fits.”<sup>14</sup> This term was first used to describe tetanus-like episodes in a young boy with a midline cerebellar tumor,<sup>15</sup> but has since been expanded to include patients with tonsillar herniation.<sup>14</sup> Pandey et al. reported that 25% of their cohort presented with such episodes, and were frequently misdiagnosed as epileptic seizures or cardiogenic syncope. However, it was also noted that decompressive surgery was sufficient to alleviate these symptoms.

Patient LG continued to be symptomatic despite surgical intervention, although the frequency of such episodes was significantly less. Data regarding surgical outcomes show variable post-operative recovery in pediatric populations. One retrospective study demonstrated symptomatic improvement in 68-72% of patients,<sup>13</sup> whereas another demonstrated improvement in 83% of patients.<sup>16</sup>

The continued occurrence of these grey spells at present remains unexplained as recent imaging did not show any obstructive pathology in the foramen magnum. Previous studies have suggested that such a recurrence may be due to the size of the decompression, post-operative scar-

ring, or re-ossification of the dura.<sup>13</sup> In addition, we hypothesize that the intrinsic neural circuitry may be inherently altered during development, and thus may not respond to surgical or pharmacological intervention. As such, we offered the family reassurance, as well as continuing surveillance and MRI.

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# A complicated presentation of peptic ulcer disease

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## CASE

Mr. SA, a 58 year-old male from London, ON, presented to the emergency department with acute intermittent right flank pain. The pain was sharp, radiated to the epigastrium, and was exacerbated with meals. A review of systems revealed no GI symptoms (including no nausea/vomiting, no change in bowel movements, no blood in stool), no constitutional symptoms, and no urinary symptoms.

Past medical history was only significant for a remote history of gastric esophageal reflux disease (GERD) or gastritis that quickly improved with one short trial of ranitidine. Routine esophagogastroduodenoscopy (EGD) and colonoscopy in 2002 was normal. Mr. SA had no prior abdominal surgery and was not on medications. He reported a 35 pack-year history of smoking and minimal alcohol intake. No family history of inflammatory bowel disease or colorectal/gastric cancer was found.

Physical exam performed by emergency staff revealed right flank pain with right costovertebral angle (CVA) tenderness as well as mild epigastric tenderness; abdominal exam was otherwise unremarkable. Blood work showed mild leukocytosis ( $11.0 \times 10^9/L$ ) and an elevated lipase (112 U/L). Liver enzymes and bilirubin were normal. Imaging with abdominal ultrasound, x-ray, and CT merely showed cholelithiasis (7mm stone) (no cholecystitis) and was otherwise normal. No renal stones were visualized. Urine dip was positive for blood. Thus, Mr. SA was diagnosed with renal colic and discharged from the emergency department with a prescription for hydromorphone.

One week later, Mr. SA returned to the emergency department with no improvement in his pain. The pain was documented as epigastric pain radiating to the right upper quadrant (RUQ) and exacerbated with meals. There was still no nausea or vomiting, no change in bowel movements, and no blood in stool. Physical exam was only notable for epigastric and RUQ tenderness. Murphy's sign was negative. On investigation, Mr. SA's bloodwork improved from the previous week: leukocyte count decreased to  $10.3 \times 10^9/L$  and lipase was normal at 27 U/L.

CT revealed new free fluid in the pelvis. There was large bowel wall thickening from the cecum to the splenic flexure with paracolic free fluid and fat stranding around the cecum and ascending colon. There was normal blood flow in the mesenteric vessels. Cholelithiasis was noted again, but this time, mild gallbladder wall thickening with pericholecystic fluid was also noted. These new findings were favoured to be reactive changes secondary to a process in the colon.

Given these findings and the unimproved abdominal pain, Mr. SA was admitted with a preliminary diagnosis of colitis (of either infectious or inflammatory etiology).

Mr. SA was made NPO and put on IV ciprofloxacin and metronidazole. He quickly became pain-free after one night in hospital. Stool ova and

parasite were negative. Culture and sensitivity could not be sent as diarrhea was not present. A colonoscopy was grossly normal from the colon to the terminal ileum. As a result, it was thought that Mr. SA likely had simple GERD or gastritis and the plan was to discharge Mr. SA following his colonoscopy with a prescription for pantoprazole.

However, on the day of expected discharge, Mr. SA developed RUQ pain. An ultrasound was ordered which revealed the following: thickened gall bladder wall (16.5mm), cholelithiasis, pericholecystic fluid, RUQ ascites, and possible microperforation of the gallbladder. Common bile ducts were normal. Radiologic impression was severe acute calculus cholecystitis.

Considering that this could be missed cholecystitis with possible perforation, Mr. SA was consented for an open cholecystectomy. In the operating room, a right subcostal incision was made. Dissection downwards revealed minimal inflammation of the gallbladder fundus. Hartmann's pouch (inferior gallbladder wall near gallbladder neck) was found to be attached adhered to the duodenum. Retracting the gallbladder revealed a punched-out hole in the duodenum and thickening in the area. No hole was found in the gallbladder.

The final diagnosis was a likely chronic perforated duodenal ulcer that sealed itself off with the gallbladder. This perforated duodenal ulcer was sealed with an omental patch (Graham patch). The gallbladder was removed and drains were left in place.

Post-operatively, Mr. SA did well and suffered no complications. A post-op small bowel follow-through showed no extravasation of contrast and no duodenal leak. Mr. SA was eventually discharged five days post-op in stable condition with a prescription for lansoprazole/amoxicillin/clarithromycin for presumed *H. pylori* infection – the likely cause of his duodenal ulcer.

## DISCUSSION

Peptic ulcer perforation is seen in 2-10% of patients with peptic ulcer disease (PUD).<sup>1</sup> Of these perforations, 60% are duodenal, 20% are antral, and 20% are gastric body.<sup>2</sup>

Classically, patients with a perforated peptic ulcer present with sudden severe and diffuse abdominal pain. Subsequently, their progression can be described in three phases.<sup>3</sup> The first phase is chemical peritonitis/contamination and occurs within 2 hours of onset of perforation as sudden epigastric pain that quickly becomes generalized. The peritoneal cavity becomes bathed with acidic fluid and the patient appears peritonitic. The second phase occurs 2 to 12 hours after onset when abdominal pain lessens. However, RLQ pain may develop as fluid moves down to the pericolic gutter. The third phase (intra-abdominal infection) presents 12-24 hours after onset. Patients notice increasing abdominal distention and become increasingly septic while abdominal pain, tenderness, and rigid-

ity may become less evident.

As described in the case, if the perforation is walled off or confined by fibrosis, symptoms may be much less severe and the three phases mentioned above may not be apparent.<sup>4</sup> Diagnosis is aided by abdominal X-ray showing free air and abdominal CT with oral contrast showing mucosal disease, inflammation, and contrast extravasation.

After initial resuscitation, intravenous proton pump inhibitors, and antibiotic coverage for gram negative species, the usual management of perforated ulcer is operative. The standard of care is closure of the duodenal defect with a piece of omentum (Graham Patch) or by truncal vagotomy with pyloroplasty. Gastric perforations can be repaired with partial gastrectomy.<sup>5</sup>

Post-operative follow-up should include upper endoscopy four to six weeks after repair to establish healing, exclude neoplasia or other diagnoses, and possibly perform biopsies for *H. pylori*.

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